PRE-SEZARY TO SEZARY ERYTHRODERMA SYNDROME

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A 50-year-old woman was admitted with pre-Se'zary syndrome in May, '97 as she had intractable progressive erythroderma with intense pruritus since 15 months. She also had axillary, inguinal, hilar and para-aortic lymphadenopathy, hepatomegaly, alopecia, hypersensitivity to cefadroxyl, and abnormal Sezary cells in peripheral blood film. Histopathology confirmed the diagnosis. Initially she was given 20mg of prednisolone which was later increased to 40mg daily but she did not respond. She responded favourably to combination of 20mg prednisolone and chlorambucil 2.5mg daily.

Key words: Se'zary cell, Erythroderma

Introduction

The pre-se'zary condition is defined as a chronic, steroid unresponsive erythroderma, lymphadenopathy, lymphocytic band at dermo-epidermal junction and fewer than 1000 circulating se'zary cells c mm.[1] Se'zary syndrome is characterised by generalised erythroderma, associated with severe pruritus, palmoplantar keratoderma, lymphadenopathy, organomegaly and circulating se'zary cells greater than 1000/c mm. In pre-se'zary, there is no abdominal organomegaly and lymphnodes reveal dermatopathic lymphadenopathy whereas in Se'zary abdominal organomegaly is present and cortical and paracortical lymphnodes are infiltrated by variable number of Se'zary cells along with abnormal lymphocytes.[2] Palmoplantar keratoderma, alopecia and onychodystrophy are progressive in Se'zary syndrome. Histopathologically lichenoid band of lymphocytes is seen in both pre Se'zary and Se'zary syndromes but in the latter abnormal lymphocytes with cerebriform hyperchromatic nuclei are seen in the dermal infiltrate, and in epidermis as focal collection of isolated/groups of cells. Dermal fibrosis is also frequent. Patients with Se'zary syndrome can have acquired monoclonal gammopathy.[3]

Case Report

A 50-year-old woman developed erythroderma since 15 months. It started as acral eczema intense pruritus of scalp and complete loss of scalp hair. The erythema and eczema progressed to back, flexures and later became generalised. Patient was admitted to PGI as a case of idiopathic erythroderma with hepatomegaly, lymphadenopathy and was treated with systemic steroids and azathioprine. She recovered, scalp hair regrew partially and was referred to contact dermatitis clinic. The disease relapsed after discontinuing the medi-

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cation and was admitted in our hospital with erythroderma and mature cataract of right eye. Skin was erythematous to hypopigmented and markedly infiltrated with leather-like feel over shins, dorsa of feet, lumbosacral region, extensors of upper limbs, lower abdomen and was interspersed with pigmented pits of variable sizes having sharp margins (Fig.1). Nails were smooth, shiny and had bevelled edges. Skin of palms and soles was thick and covered with scales. She developed hypersensitivity to cefadroxyl during admission. Two years back, she was operated for cataract of left eye. Multiple, 1.5 to 3cm, freely mobile, discrete, non-tender, firm, lymphnodes were palpable in the axillae and groins. Liver was enlarged but nontender. Investigations revealed Hb-8.5%, TLC-8400/cmm, DLC-P56, L21, E20, M2, B1, and platelet count 3.7 lakh/cmm. Urinalysis was normal. FBS-72mg% SGOT-18 IU/L, SGPT-20 IU/L. Peripheral blood film revealed atypical lympocytes with cerebriform nucleus i.e, Se'zary cells (Fig.2). X-ray chest showed hilar and middle mediastinal lymphadenopathy. USG abdomen revealed multiple hypoechoic areas in

Fig. 1. Hypopigmented, markedly infiltrated skin covered with scales and interspersed with pigmented pits having sharp margins.

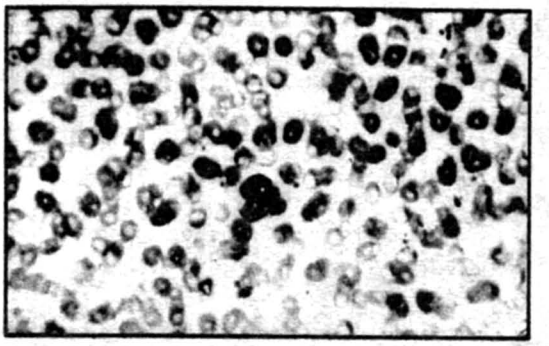

Fig. 2. Cerebriform Se'zary cell in peripheral blood film. (Leishman's stain)

right lobe of liver and para-aortic lymphadenopathy. FNAC from right inguinal lymphnodes showed reactive hyperplasia while biopsy was of dermatopathic lymphadenopathy. Skin biopsy on both admissions revealed hyperkeratosis, keratotic plugging, acanthosis, prolongation of rete ridges, papillomatosis, dermal mononuclear infiltrate with sharp lower border and some abnormal lymphocytes. In second biopsy mononuclears were admixed with se'zary cells and some of the cells infiltrated the epidermis. Dermal fibrosis was prominent at one area.

Discussion

Present case was diagnosed initially as pre-Se'zary syndrome because of intractable erythroderma, intense pruritus, hepatomegaly, convolution of skin of forehead, widespread lymphadenopathy and Se'zary cells in peripheral blood smears. She did not respond to even 40 mg prednisolone daily and se'zary cells increased in peripheral blood and dermal infiltrate as evolution into se'zary syndrome occurred. Favourable response to combined therapy of 20 mg prednisolone and 2.5mg chlorambucil was visible within first week of therapy.

Early diagnosis and treatment of se'zary and pre-se'zary syndromes is rewarding.

References

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